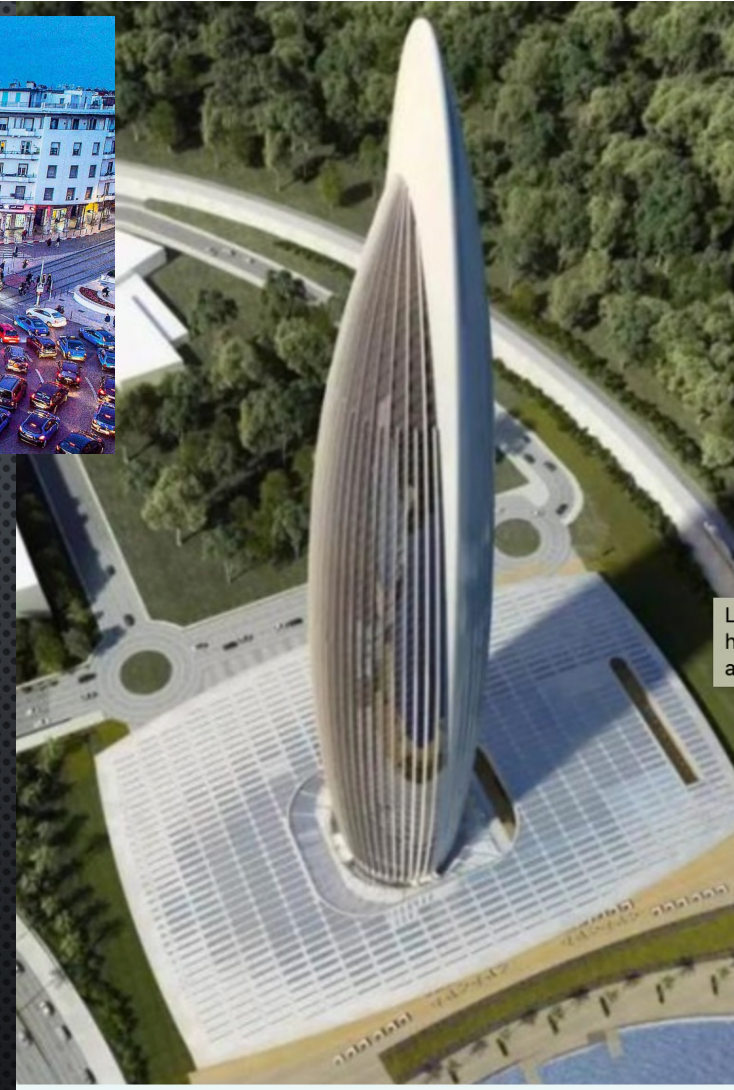


Rabat city MOROCCO



OHVIRA SYNDROM

PR. JAMAL FIKRI

- **REPRODUCTIVE AND SURGERY AND ART SPECIALIST**
- HEAD OF POSTGRADUATE DEGREE ON REPRODUCTIVE MEDECINE
- HEAD OF ART UNIT AL BOUSTANE - RABAT-MOROCCO
- **CHAIRMAN OF MOROCCAN COLLEGE OF FERTILITY**



INTRODUCTION

- MULLERIAN ANOMALIES ORIGINATE FROM **ADVERSE EMBRYOLOGIC EVENTES** AND PRESENTS IN SEVERAL AND VARIOUS WAY RANGING FROM INCOMPLETE AGENESIS TO LATERAL AND VERTICAL FUSION DEFECT
- OHVIRA SYNDROME IS THE RESULT OF AN EMBRYOLOGICAL ARREST OF THE MÜLLERIAN AND MESONEPHRIC DUCTS AT 8-9 WEEKS OF GESTATION

INTRODUCTION

- THE EXTENSIVE USE OF **ULTRASOUND** AND OTHER IMAGING MODALITIES ALLOW TO DISCOVER MORE AND MORE MALFORMATIONS BEFORE ANY CLINICAL MANIFESTATION, RISING DIFFICULTIES ON HOW TO INFORM PATIENTS AND MANAGE THIS CONDITIONS, THAT **SOME WILL NEVER SHOW ANY CLINICAL MANIFESTATION**
- **NON-OBSTRUCTIVE ANOMALIES** ARE THE MOST FREQUENT AND MOSTLY DIAGNOSED DURING ADVERSE OBSTETRICAL OUTCOMES OR FERTILITY CONCERNS
- **OBSTRUCTIVE MALFORMATIONS** EXPRESS THEMSELVES AT THE PUBERTY WITH CYCLIC PERLVIC PAIN ASSOCIATED TO AMENORRHEA OR PELVIC MASSE RELATED TO MENSTRUAL RETENTION.

INTRODUCTION

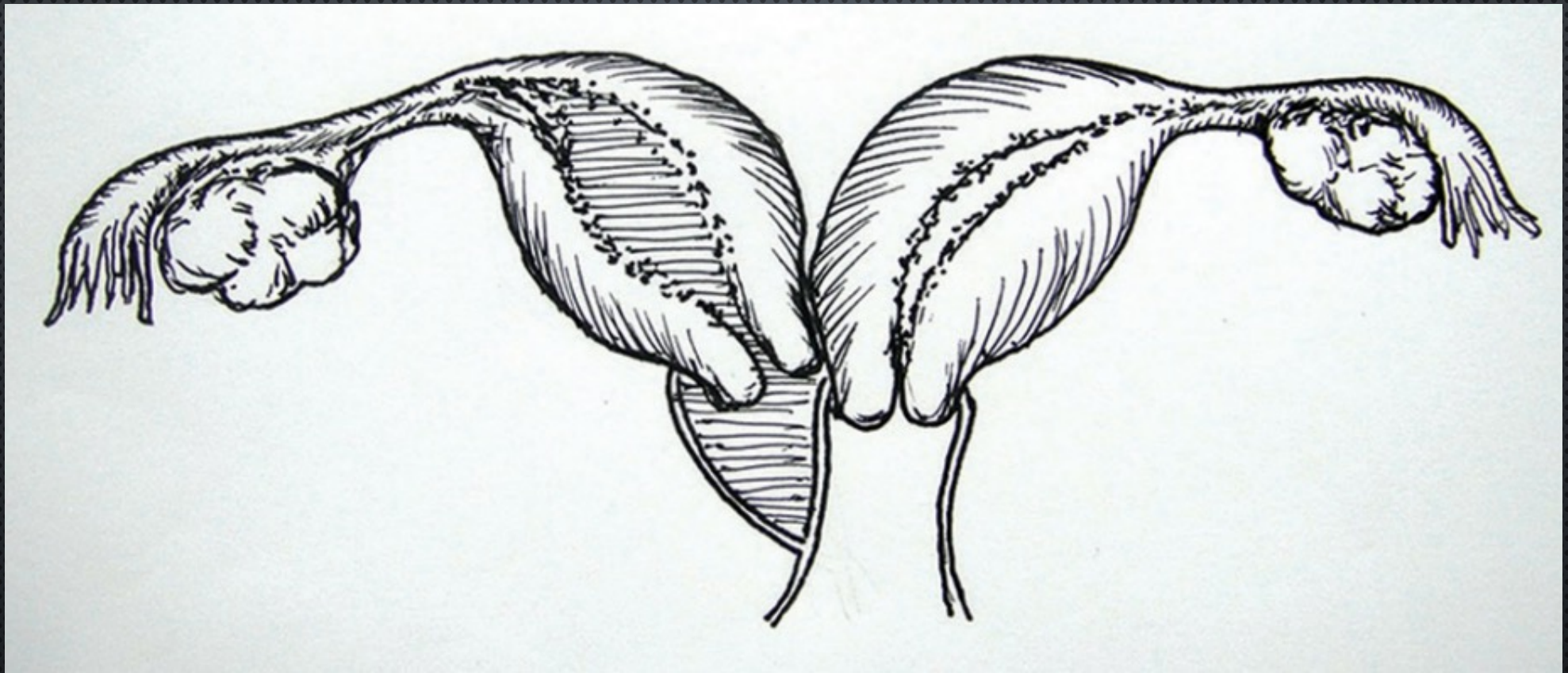
- DUPLICATED UTERUS WITH BLIND HEMIVAGINA PRESENTING WITH HEMATOCOLPOS, HEMATOMETRA AND HEMATOSALPINX WAS DESCRIBED FIRST IN 1922.
- 1976 SOME CASES ASSOCIATING VARIOUS ASSOCIATIONS LEADS TO DESCRIBE THE
- HERLYN-WERNER-WUNDERLINCH SYNDROME
- TRIADE :

DUPLICATED UTERUS

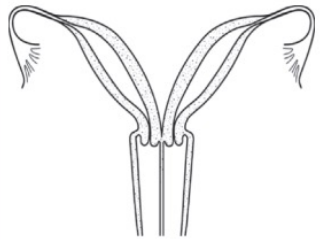
OBLIQUE VAGINAL SEPTUM OBSTRUCTING MENSES OUTFLOW

RENAL AGENESIS

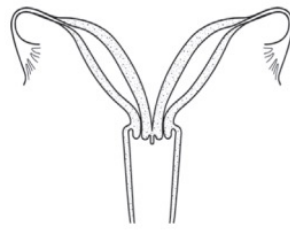
- SMITH AND LAUFER INTRODUCED THE CONCEPT OF **OHVIRA** (**OBSTRUCTION** + **RENAL ANOMALIES**)



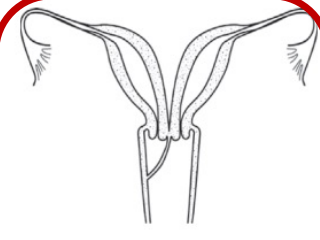
UTERUS DIDELPHYS



UTERUS DIDEPHYS AND
LONGITUDINAL SEPTUM

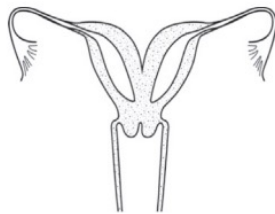


UTERUS DIDELPHYS AND
+/- LONGITUDINAL VAGINAL
SEPTUM OF VARIABLE LENGTH

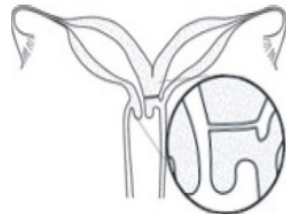


UTERUS DIDELPHYS AND
OBSTRUCTED R/L HEMIVAGINA

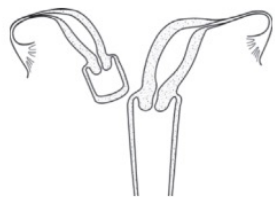
COMPLEX ANOMALIES



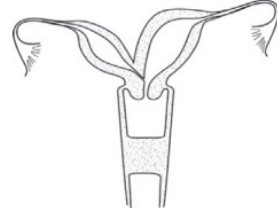
BICORNUATE UTERUS WITH
BILATERAL OBSTRUCTED
ENDOMETRIAL CAVITIES



UTERUS DIDELPHYS WITH
COMMUNICATING HEMIUTERI AND
UNILATERAL R/L CERVICO-VAGINAL
ATRESIA



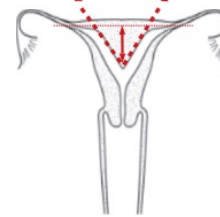
OBSTRUCTED R/L HEMIVAGINA,
HEMIUTERUS AND SINGLE CERVIX
WITH SEPARATE CONTRALATERAL
R/L PATENT HEMIUTERUS, CERVIX
AND VAGINA



BICORNUATE UTERUS WITH
R/L COMMUNICATING TRACT AND
TRANSVERSE VAGINAL SEPTUM

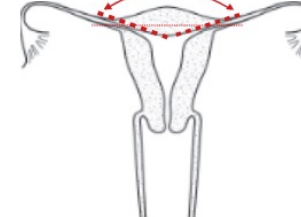
SEPTATE UTERUS

Septum length >1 cm
Septum angle <90°

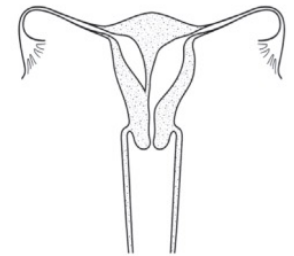


PARTIAL SEPTATE UTERUS

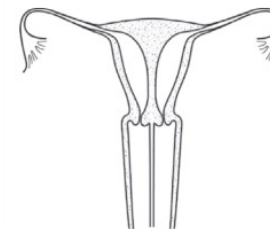
Arcuate/Normal
length ≤1cm angle >90°



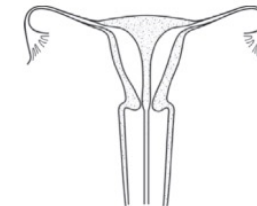
NORMAL/ARCUATE UTERUS



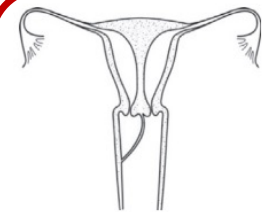
ROBERT'S UTERUS



COMPLETE SEPTATE UTERUS
WITH DUPLICATED CERVICES AND
LONGITUDINAL VAGINAL SEPTUM



COMPLETE SEPTATE UTERUS WITH
SEPTATE CERVIX AND
LONGITUDINAL VAGINAL SEPTUM



COMPLETE SEPTATE UTERUS,
DUPLICATED CERVICES, AND
OBSTRUCTED R/L HEMIVAGINA

ASRM müllerian anomalies classification 2021



ESHRE/ESGE classification Female genital tract anomalies



Uterine anomaly			Cervical / Vaginal anomaly	
	Main class	Sub-class	Co-existent class	
U0	Normal uterus		C0	Normal cervix
U1	Dysmorphic uterus	a. T-shaped b. Infantilis c. Others	C1	Septate cervix
U2	Septate uterus	a. Partial b. Complete	C2	Double "normal" cervix
U3	Bicorporeal uterus	a. Partial b. Complete c. Bicorporeal septate	C3	Unilateral cervical aplasia
U4	Hemi-uterus	a. With rudimentary cavity (communicating or not horn) b. Without rudimentary cavity (horn without cavity / no horn)	C4	Cervical Aplasia
U5	Aplastic	a. With rudimentary cavity (bi- or unilateral horn) b. Without rudimentary cavity (bi- or unilateral uterine remnants / Aplasia)	V0	Normal vagina
			V1	Longitudinal non-obstructing vaginal septum
			V2	Longitudinal obstructing vaginal septum
			V3	Transverse vaginal septum and/or imperforate hymen
			V4	Vaginal aplasia
U6	Unclassified Malformations			
U			C	V

New Classification of Herlyn-Werner-Wunderlich Syndrome

Lan Zhu, Na Chen, Jia-Li Tong, Wei Wang, Lei Zhang, Jing-He Lang



Figure 1: Classification 1.1, with blind hemivagina.

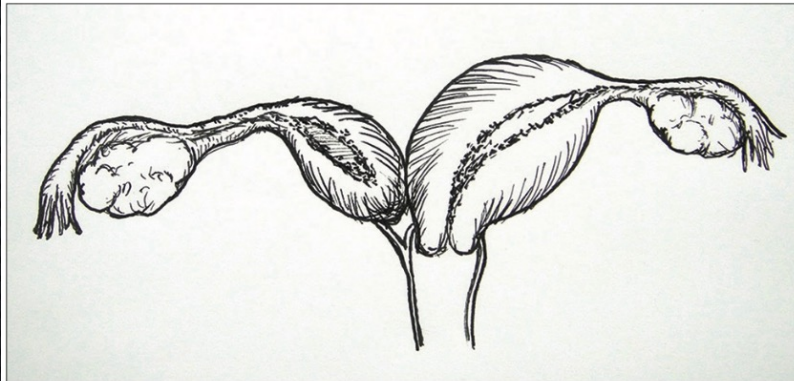


Figure 2: Classification 1.2, cervicovaginal atresia, without communicating uteri.



Figure 3: Classification 2.1, partial reabsorption of the vaginal septum.



Figure 4: Classification 2.2, incompletely obstructed hemivagina with communicating uteri.

SYMPTOMS

- **SYMPTOMS APPEARS VERY EARLY AFTER MENARCHE**
- DYSMENORRHEA WITH PROGRESSIVE WORSENING
- PELVIC PAIN
- PELVIC TENDER MASS
- UROLOGICAL SIGNS RELATED TO COMPRESSION
- VAGINAL FOUL DISCHARGES (INFECTION OF THE COLLECTION)



Three-dimensional ultrasound in the diagnosis and the classification of congenital uterine anomalies using the ESHRE/ESGE classification: a diagnostic accuracy study

Anna Kougioumtsidou¹ · Themistoklis Mikos¹ · Grigoris F. Grimbizis¹ · Aikaterini Karavida¹ · Theodoros D. Theodoridis¹ · Alexandros Sotiriadis¹ · Basil C. Tarlatzis¹ · Apostolos P. Athanasiadis¹

Ludwin and All; JMIG 2013
Similare findings

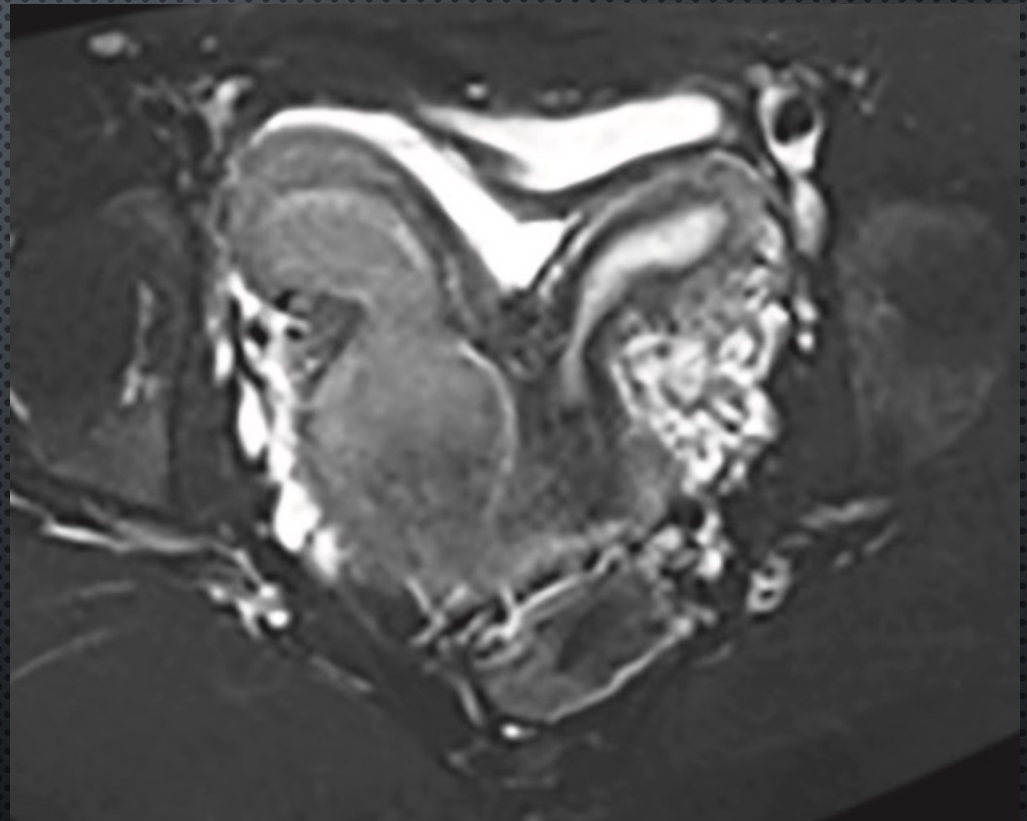
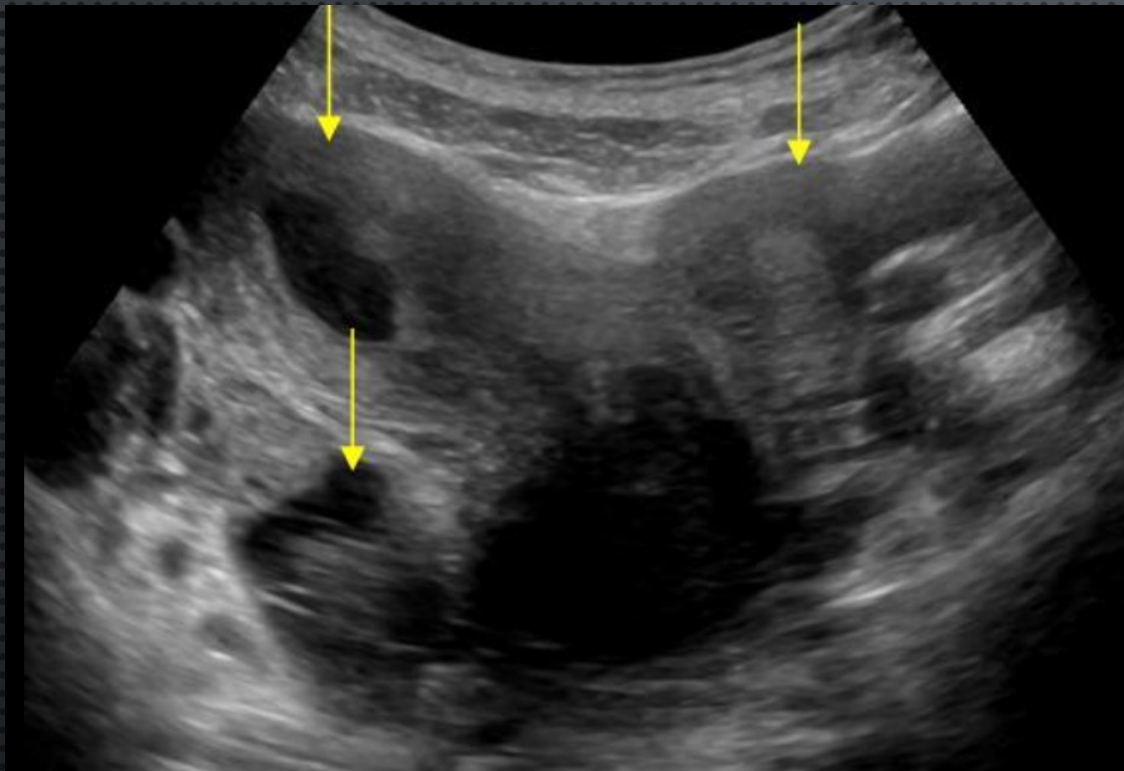
Septate uteri:

Sensitivity 100%, Specificity 92.3%, PPV: 98%, NPV: 100%

Diagnostic accuracy = 100% for
aplastic, dysmorphic uteri unicornuate and bicornuate

PRACTICE COMMITTEE OF THE AMERICAN SOCIETY FOR REPRODUCTIVE MEDICINE, 2016

- NON INVASIVE DIAGNOSTIC MODALITIES ARE RECOMMENDED AS GOLD STANDARD
- **3D TVUS IS THE BEST AND MOST ACCURRATE TOOL**
- HSG, **MRI**, LAPAROSCOPY AND HYSTEROSCOPY FOR COMPLEXE CASES AND CONCURRENT PATHOLOGIES SUCH AS ENDOMETRIOSIS ETC.....

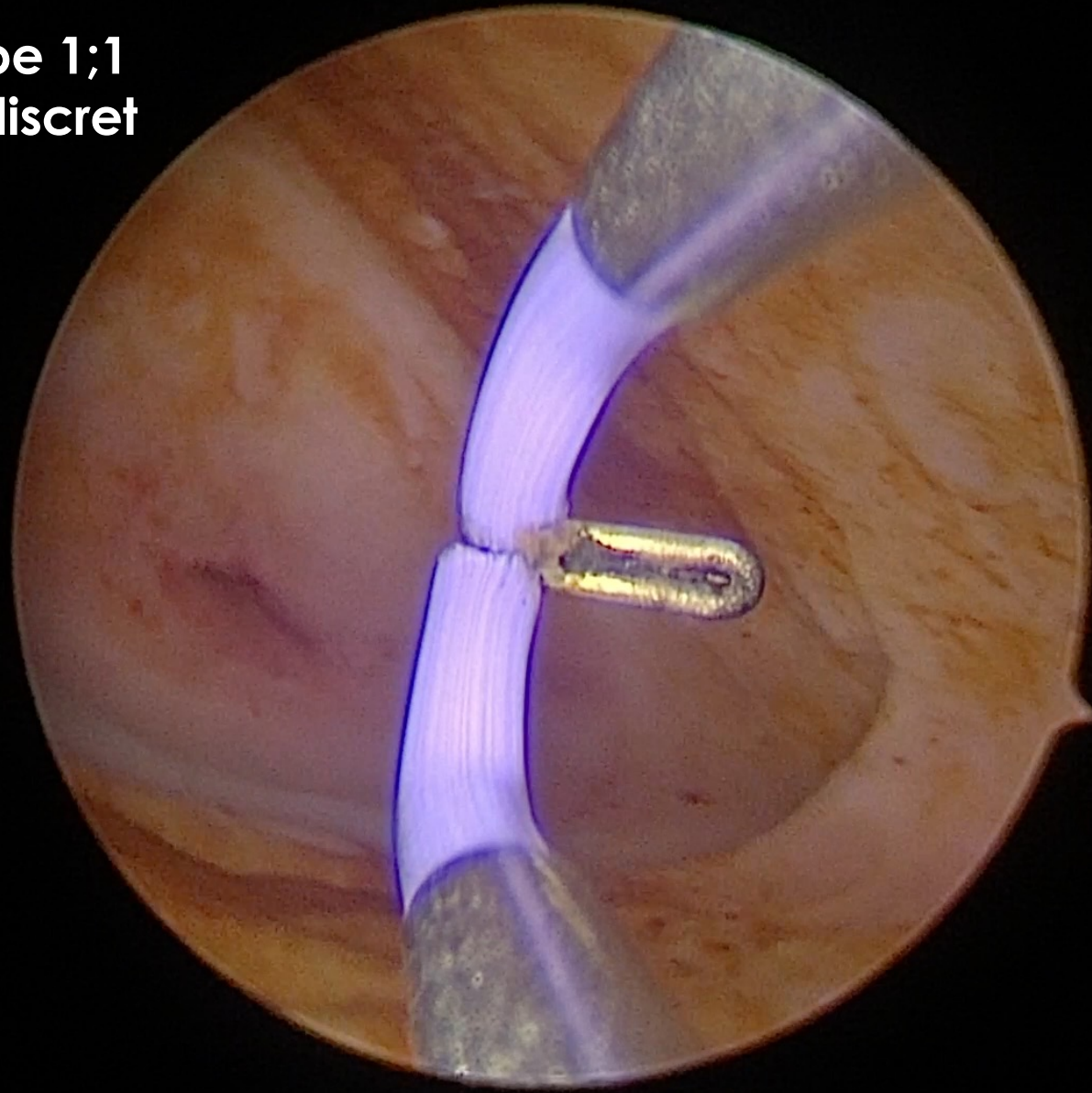


- **VAGINAL SEPTOPLASTY IS THE MAINSTAY SURGICAL APPROACH**
- **VAGINOSCOPIC/HYSTEROSCOPIC BIPOLAR RESECTION IS THE ONLY WAY FOR VIRGO**

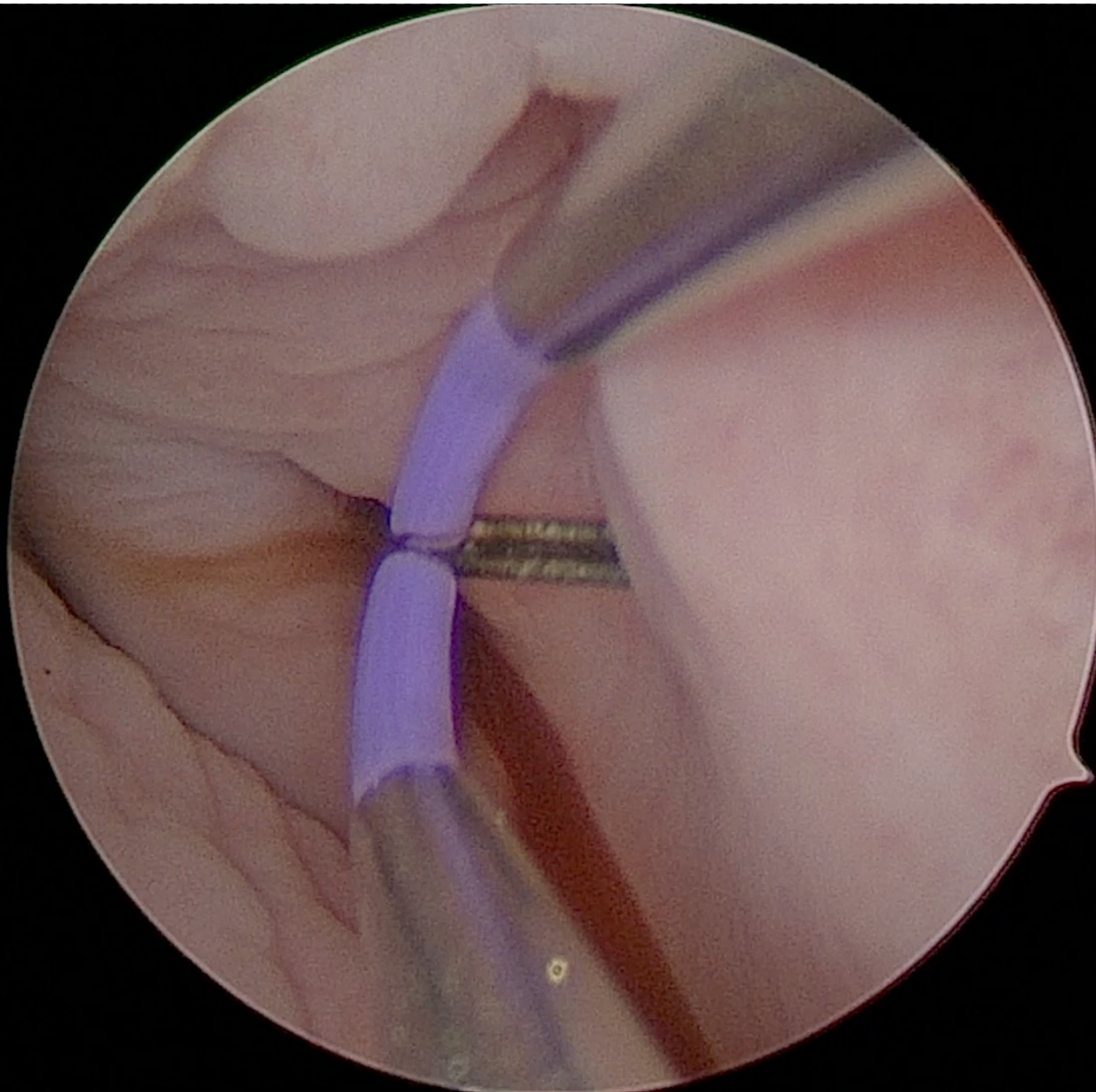


Cas. 1
Type 1:1

Cas : 2 et 3 . Type 1;1
Hematocolpos discret



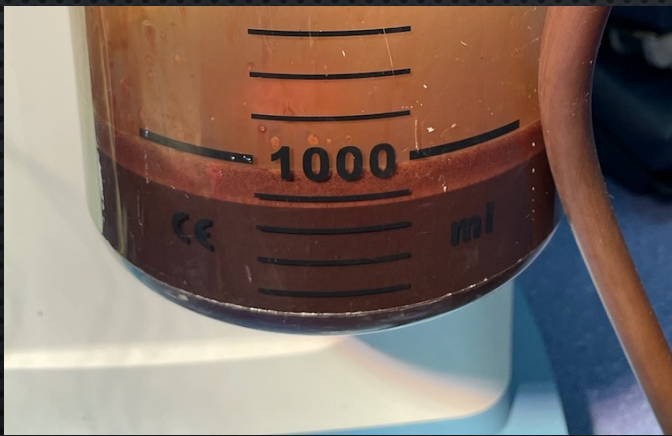
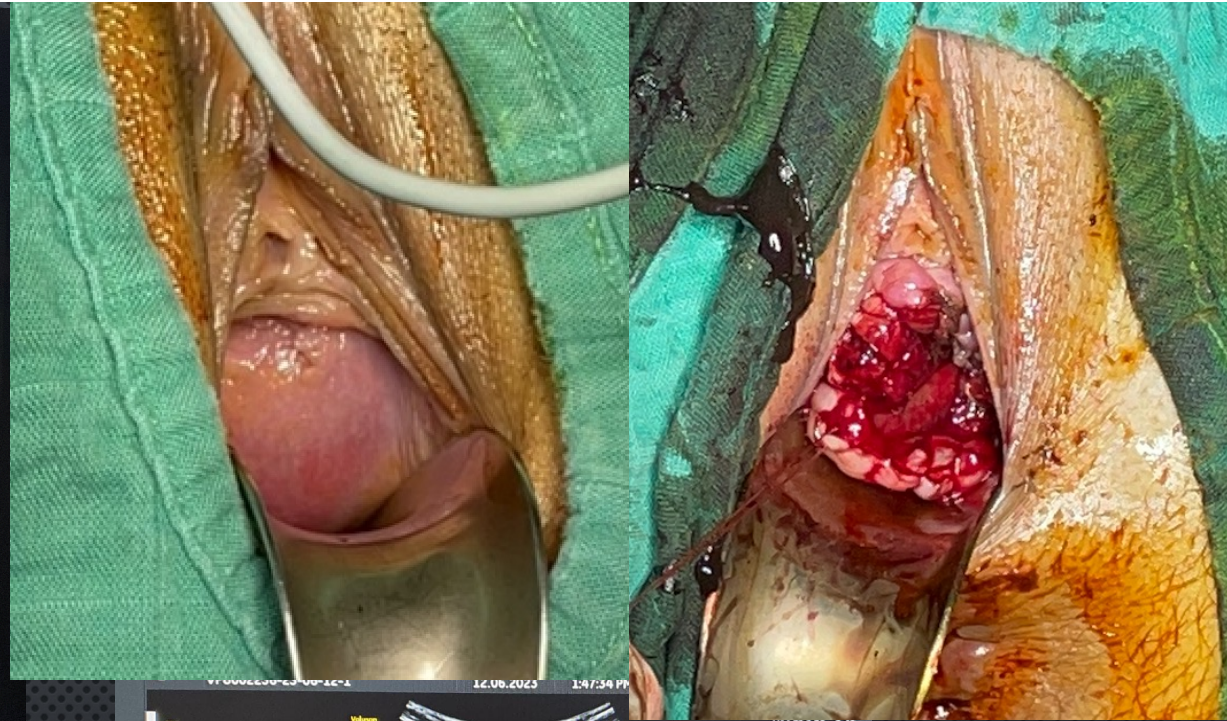
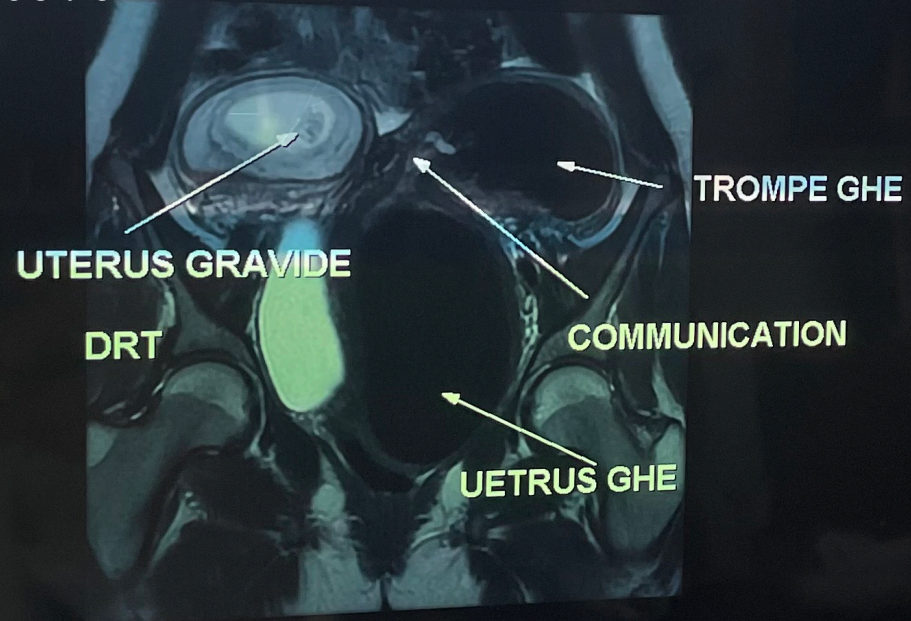
Cas : 4
Type 2;2



CAS : 5 PRISE EN CHARGE INADÉQUATE

- 14 ANS , PUBERTÉ DE PUIS 1 AN
- DYSMÉNORRHÉES INVALIDANTES AVEC AGGRAVATION PROGRESSIVE
- ECHOGRAPHIE PARLE DE MALFORMATION UTERINE AVEC HEMATCOLPOS
- PONCTION VAGINALE SIMPLE
- **ABCÈS TUBAIRE >>> SALPINGECTOMIE LAPROTOMIQUE**
- MISE SOUS PILULE EN CONTINU POUR BLOQUER LES RÉGLES. >>> ARRÊT PILULE
- REPRISE DES DOULEURS
- TRAITEMENT HYSTEROSCOPIQUE AVEC ÉVOLUTION FAVORABLE
- **PERTE D'UNE TROMPE ET PROBABLEMENT ATTEINTE DE LA TROMPE RESTANTE**

Cas : 6



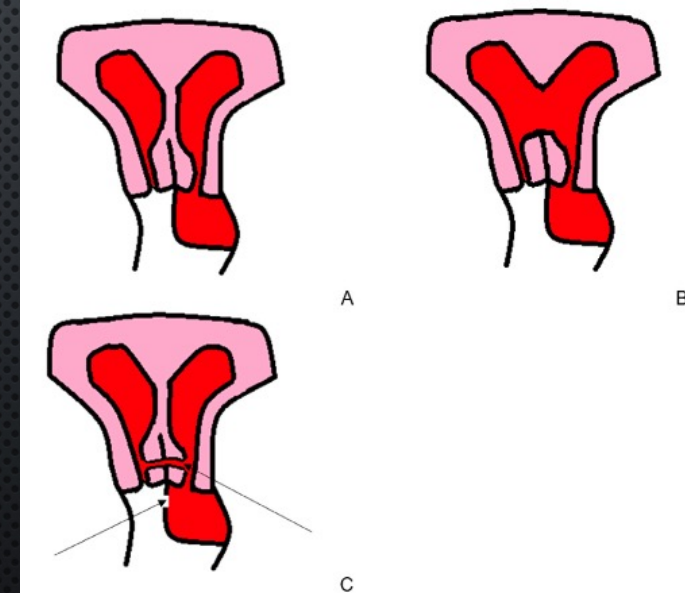
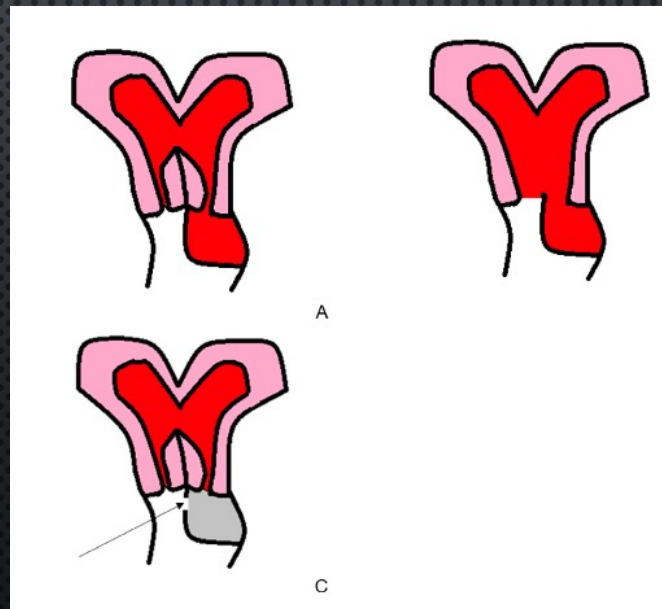
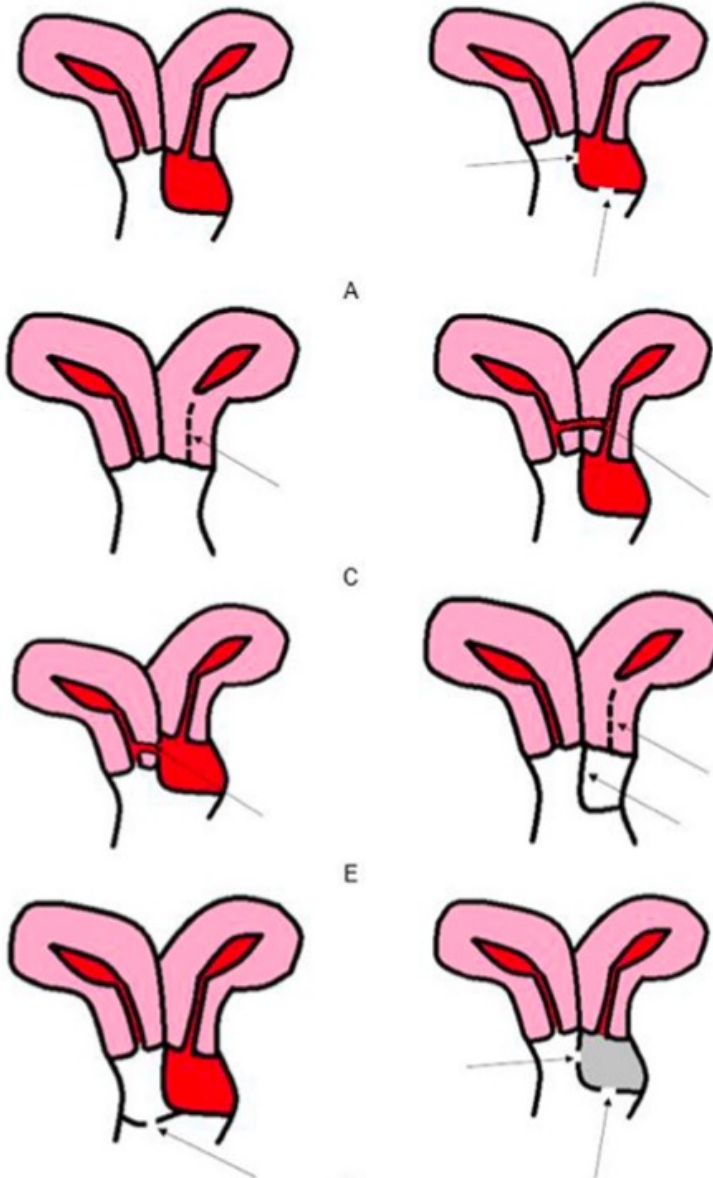
Herlyn-Werner-Wunderlich syndrome also known as obstructed hemivagina and ipsilateral renal anomaly: A case report and a comprehensive review of literature

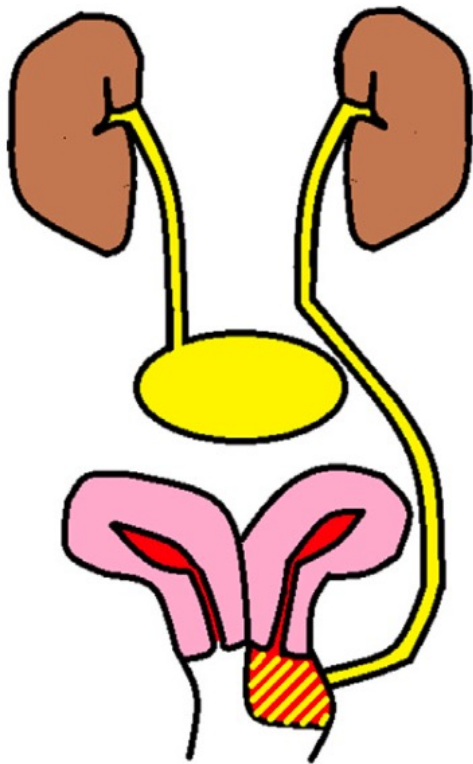
Radiology case reports 2023

André Luís Borges, MD^{a,b,*}, Nhalim Sanha, MD^a, Helena Pereira, MD^a, Ana Martins, MD^a, Cristina Costa, MD^a

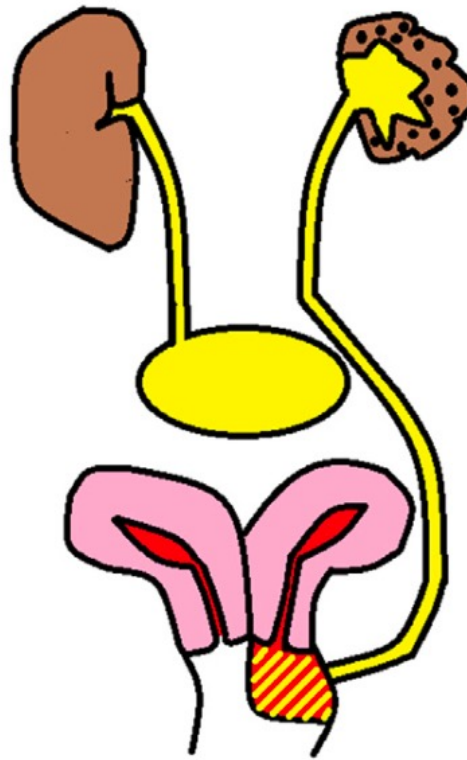
Table 3 – Types of reported Mullerian anomalies according to the ARSM classification.

Uterus Didelphys	462 (88,7%)
Bicornuate Uterus	34 (6,5%)
Septate Uterus	25 (4,8%)





Ureteric reimplantation



**Hemi-hysterectomy
+ Uretero-nephrectomy**



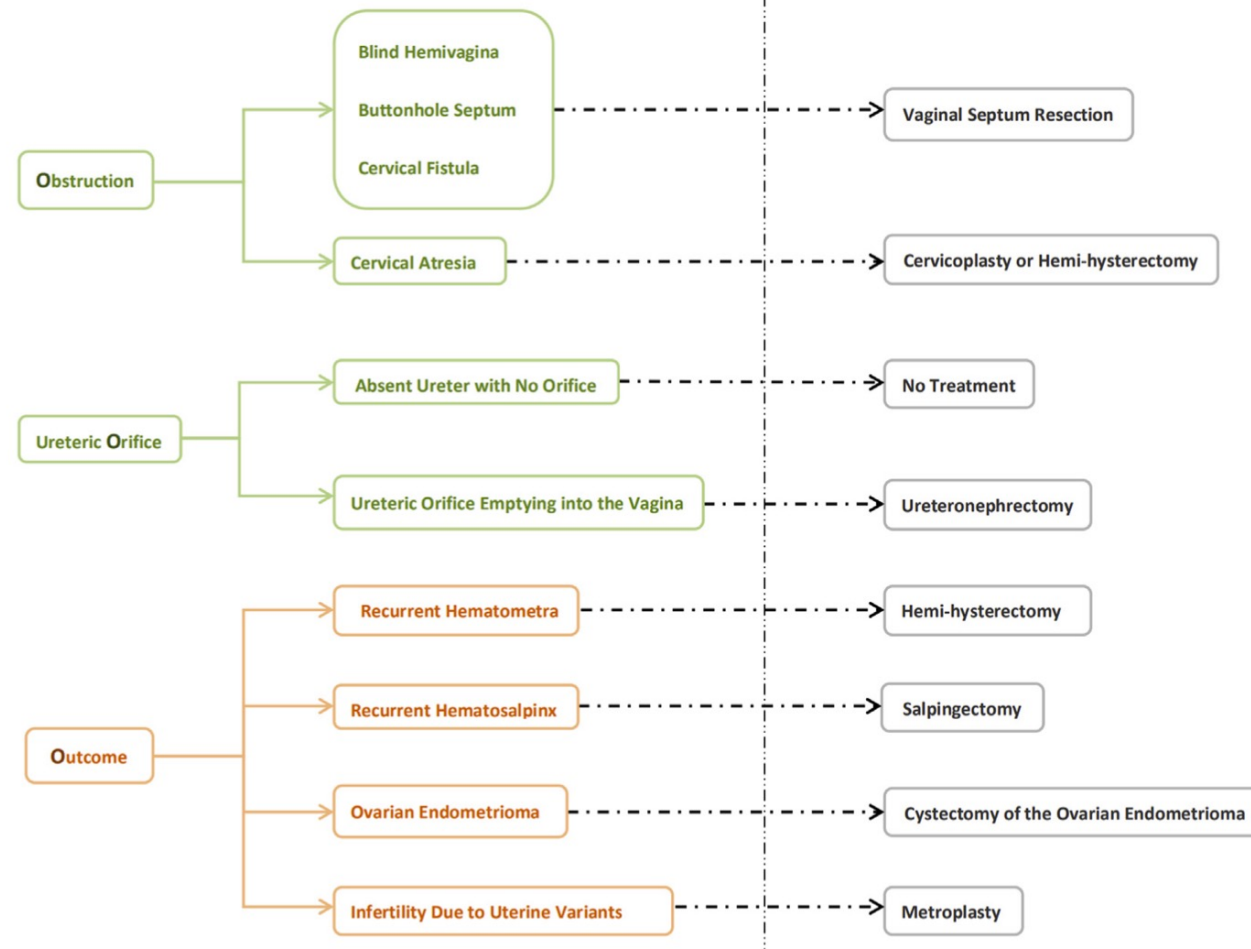
BICORNUATE (CL.4 AFS) AND DIDELPHYS UTERI (CL 3 AFS) (CLASS U3 ESGE). (FUSION DEFECT)

- 10 % OF ALL UTERINE ANOMALIES AND **1/1000** WOMEN OF REPRODUCTIVE AGE
- **1/5 DUPLICATED UTERI IS OHVIRA**
- DIDELPHIC UTERI SEEMS TO HAVE THE BEST REPRODUCTIVE OUTCOMES THAN OTHER UTERINE MALFORMATIONS OR AT LEAST SIMILAR TO THOSE OF UNICORNUATE UTERI.
- 94 % OVERALL PREGNANCY RATE
- 21 % MISCARRIAGE RATE
- **30 % PRETERM BIRTH**
- THE IMPLANTATION RATE IS ESTIMATED TO BE LOW (5.9 %)
- **PREGNANCY RATE : 11.1 %**

HEINONEN P. EUROPEAN JOURNAL OF OBSTETRICS & GYNECOLOGY AND REPRODUCTIVE BIOLOGY (2000)

Proposal of the 3O (Obstruction, Ureteric Orifice, and Outcome) Subclassification System Associated with Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) 2020

Junjie Zhang MD¹, Mengda Zhang MD², Yu Zhang MD³, Huining Liu MD³, Peng Yuan MD¹, Xiaoyan Peng MD⁴, Zhenzhen Cao

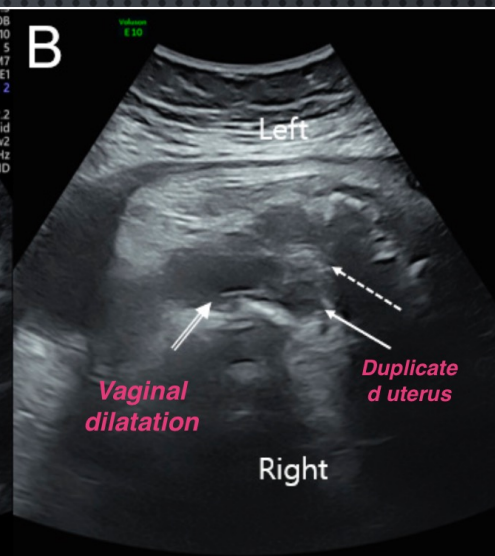
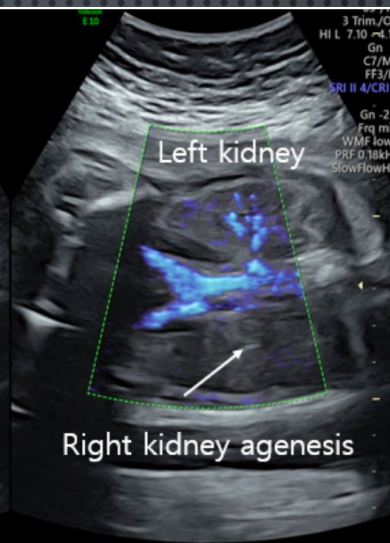
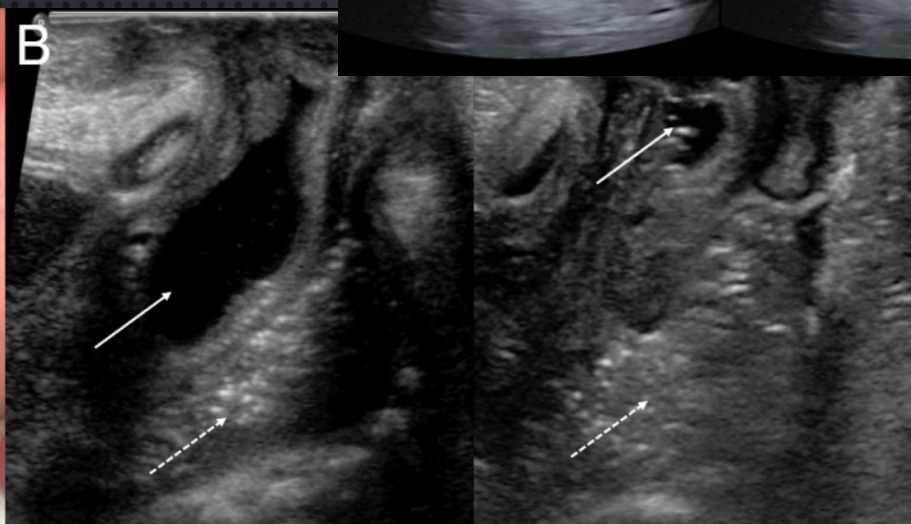
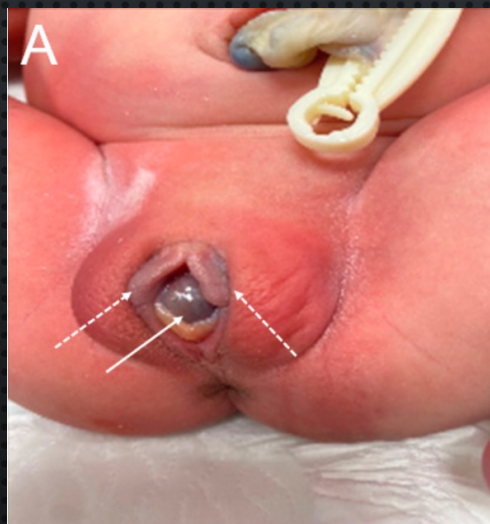




Case Report

Prenatal Diagnosis of Fetal Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) Syndrome

Soo Jung Kim ¹ , So-Yeon Shim ², Hyun-Hae Cho ³, Mi-Hye Park ¹ and Kyung A. Lee ^{1,*}



PRACTICAL POINTS

- DYSMENORRHEA RAPIDLY WORSENING AT THE MENARCHE SHOULD PROMPT US WORK UP
- OBSTRUCTIVE ANOMALY SUSPECTED : PRECISE DIAGNOSIS IS MANDATORY BEFORE ANY HASTY SURGERY
- CLEAR RECOGNITION OF RENAL ANOMALIES TO PERFORM THE PROPER SURGERY
- PRENATAL DIAGNOSIS OF FOETAL KIDNEY ANOMALY IS NOT DIFFICULT AND SHOULD DRAW ATTENTION TO POSSIBLE GENITAL TRACT MALFORMATION
- ACCURATE KIDNEY FUNCTION IF ANY IS TO BE PERFORMED VERY SOON AT THE CHILDHOOD IN AN ATTEMPT TO SAVE THE ORGAN IF POSSIBLE
- EARLY TREATMENT OF THE OBSTRUCTIVE ANOMALIES COULD PREVENT COMPLICATIONS SUCH AS ENDOMETRIOSIS AND PELVIC INFECTIONS THAT NEGATIVELY IMPACT REPRODUCTIVE POTENTIAL